THOMSEN'S DISEASE—REPORT OF A CASE.[1]

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THOMSEN'S disease, or congenital myotonia, is so rare a malady that the report of an additional case will, I venture to believe, prove of interest. Most of the cases recorded have occurred in Germany, and, curiously, in the present instance the patient was a German by birth, although he migrated to this country in early boyhood.

In March last, by request of Dr. J. W. Whitbeck, of Rochester, August B—— consulted me for the peculiar symptoms so significant of this disorder. The patient was twenty-one years of age, single, and for the past five years had followed the trade of a clothing cutter. His general health was excellent and the only noteworthy illness had been an ordinary attack of typhoid fever at the age of five. The family history was negative, with one important exception. His oldest brother had suffered from the same affection twenty-five years ago, and a few months prior to the birth of my own patient, shot himself to avoid constant penalty and disgrace. Compelled to enter the German army, and unable to execute his duties, he was constantly subjected to the harsh discipline awarded malingering, since at that time the disease had not been recognized, and he resorted to the only way of escape. In the present case the disease was congenital, the characteristic symptoms appearing in infancy and gradually becoming more marked, though not till two years ago did they cause serious inconvenience. The interference with immediate muscle action became pronounced and he ascended stairs or arose from a chair with difficulty. Persistent contracture of the calf muscles developed sufficiently to cause a permanent talipes equinus, due perhaps to the demands of his work, which required him to stand the whole day. So powerful was the spasm of the affected muscles that a slight push or slip would result in a fall, the least involuntary effort to recover himself proving disastrous. Truly he was being mastered by his own muscles.

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At the time he consulted me this muscular abnormality was well developed. The muscles were hard and prominent, athletic in appearance, although unsymmetrically enlarged. In normal muscular development the muscle mass is symmetrical throughout. In this pathological condition the belly of the muscle is over-developed, producing a peculiar deformity, differing, I believe, very materially from pseudo-muscular hypertrophy, which disease it superficially resembles. Voluntary effort of the affected muscles after a period of repose immediately developed tonic spasm, momentarily inhibiting the intended action. The rigidity then slowly relaxed, and when once the spell was broken, normal control was easily sustained. The peculiar "intentions krämpfe" were most marked when the patient undertook to regain his feet after a fall. His efforts to use the muscles, rigid at the initial impulse, were violent but fruitless, and only by climbing up his legs with the aid of his hands, as in pseudo-muscular hypertrophy, could he accomplish his purpose. The muscles affected were the quadriceps extensor of the thigh, the calf muscles, the deltoid, and, to a lesser degree, the others of the legs, arms and trunk. The muscles of the neck, face and head were unaffected, and there was no interference with speech or with swallowing. He had, however, noted some difficulty in relaxing the grasp of the hand.

Mechanical stimulation of the muscles produced energetic tonic contractions, succeeded by gradual relaxation, causing the dimpling and furrowing generally observed. My experience in the present case was that this reaction was most readily obtained by percussion upon the tendinous insertions of the muscles. Under continued mechanical stimulation the reaction became weaker and was ultimately lost.

Galvanic stimulation gave the characteristic reactions, but even with a current of 20 or 30 milliampères the peculiar undulatory contractions described by Erb could not be satisfactorily obtained, although rythmic contractions were noted during the period of circuit closure. The contractions were sudden, persistent and painful, continuing for twenty or thirty seconds after removal of the electrodes. The galvanic formula was as follows, both normal and abnormal reactions being given for the sake of comparison :

Gal. Amp. .020. Electrode disk .005 m.	Thigh Muscles (Patient).	Arm Muscles (Self).
K. C. C.	100	20
A. C. C.	50	10
A. O. C.	10	0
K. O. C.	0	0

The formula indicates increased galvanic irritability, but no qualitative change other than a distinct anodal opening contraction. Response of the muscles to faradic stimulation was pronounced, prolonged and very painful, while the anodal contraction was even more marked than the kathodal. The same dimples and furrows obtained by mechanical excitation were developed, and were even more persistent after cessation of the current. The reaction of the nerves to either current was unaltered.

All the reflexes, superficial and deep, were intensified; the knee-jerk especially marked and much increased by "re-enforcement." There was no sensory disturbance of any sort, all forms of stimulation giving normal response. The dynamometer, recently perfected by Tiemann, indicated a surprising weakness even in the forearm muscles not materially affected. The grip gave a pressure of 110 pounds for the right hand and 80 pounds for the left, about a third of the normal rate.

One reaction was obtained which has not, to my knowledge, yet been noted. A decided pin prick produced a clean cut, punch like perforation, surrounded by a distinct white wheal. So prolonged was the resulting muscle spasm that, although the pin was thrust completely through the skin, bleeding did not occur for several minutes. In no other myopathy have I noted so marked a reaction of this sort, and I suggest it as an additional diagnostic sign. Fibrillar twitchings and the double contraction under percussion, noted in progressive muscular atrophy, were never observed.

The ocular examination was kindly made by Dr. Sumner Hayward, of Rochester, with the following result: Right eye normal, save a few opacities in vitreous and myopia one dioptre; left eye normal in every way. Pupillary reaction normal, no contraction of field, no dyschromatopsia; relative movements of lids normal. The eye, when made to follow quickly the movements of a pencil in all directions, revealed a jerky indecision, not promptly following the pencil and then not fixing accurately but, as it were, seeking the pencil before becoming steady; muscular balance of the eyes perfect.

The extremities were cold and bluish in tint, from impeded circulation. No microscopical examination of the muscular fibre was attempted.

In the treatment of this case two or three points are worthy of mention. When he first consulted me, galvanism with a strength of amp. .020 or .025 was employed alternate days with very meagre results, and shortly its use was discontinued. Later the bromide of sodium was given in excessive doses, with an ill-defined idea that it might lessen the muscular stiffness. For three days the patient took fully an ounce of the salt daily, in four divided doses, with a somewhat unexpected result. It did reduce muscular activity, but apparently in the less affected muscles only, intensifying seriously the muscular disturbance.

The muscles which responded normally to voluntary impulse were much weakened, while those subject to the disorder were not materially influenced by the drug. Locomotion was almost impossible, the patient stumbling over his own feet, unable to get them out of his way; while the cramping of the affected muscles was painfully aggravated. The usual symptoms due to bromide intoxication were well marked, while all the mechanical and electrical reactions were somewhat lessened. The ill effects of the drug passed off within a few days, leaving my patient neither better nor worse for the experiment. I only refer to this in passing, because of the bearing this therapeutical test may have upon the essential nature of the disease. Certainly the excessive response to reflex stimulation, the difficulty of establishing voluntary control, as well as the marked variation in the muscular phenomena under the influence of the depressor motor drug, point rather conclusively to the local nature of the disorder.

Knowing the hopelessness of all ordinary methods, we resorted to the following expedient, assured that whatever might be the remote cause of the disease, excessive muscular irritability was at least its expression. The patient was put in bed, and the ordinary rubber bandage was applied to the legs, leaving the knees free. Our purpose was to limit the activity of the affected muscles and produce sweating, hoping thereby to secure relaxation and perhaps retrograde metamorphosis. At least the elastic bandage gave support and resulted in very material improvement. The ordinary cotton roller, employed for a time, was unsatisfactory and as the rubber bandage was troublesome, gum sheeting, the thickest possible, was substituted. An elastic corset was carefully fitted to either calf and thigh and made with reenforced eyelets along either margin, so that it could be laced tightly about the part. By this elastic covering the muscles of both legs were firmly bound and very satisfactorily supported. Steady improvement in his gait and equipoise was noted, while the stiffness and pain were much abated. Unfortunately, circumstances would not admit of any prolonged stay at the hospital, and two weeks after the application of the rubber supports he was allowed to go home and directed to continue their use. At this time he no longer fell, on stumbling; walked easily, without the usual struggle to get started, and in most ways could very well control muscle action. However, the inherent muscular weakness was unaltered and the reactions but slightly modified. Unfortunately, no pure rubber sheathing of sufficient thickness could be obtained to resist the constant strain from exercise and an ordinary elastic stocking was substituted. This, however, did not answer the purpose, nor did a pure rubber facing make the stockinet more efficient, and some six or eight weeks after his return home all the difficulty in gait returned. However, I am satisfied that a sheathing of pure rubber gum, with limitation of muscular exertion for an extended period, will very markedly ameliorate the symptoms, at least in this case. Could the rest in bed have been sufficiently prolonged, I am confident the marked improvement would have been more satisfactory.

DIAGNOSIS OF LESIONS OF THE OPTIC LOBES.

Eisenlohr reports the case of a young man, twenty-eight years of age, who was struck in the head with a bullet from a revolver, the ball being found in the tubercula quadrigemina at the necropsy, who developed the following symptoms :

1. Tremor of the left arm, with unequal pupils, following secondary irritation of the pyramidal tract.
2. Associated paralysis of the eye-muscles, with diminution of the acuteness of vision, this latter symptom being due to a lesion of the optic lobes, the ophthalmoplegia depending upon a concomitant lesion of the nucleus of the occulo-motorius.
3. Polyuria without glycosuria, cerebellar ataxy, and finally symptoms of cerebral compression. The writer emphasizes the existence of visual and pupillary disturbances in lesions of the optic lobes.—(Rivista clinica e Terapeutica, 15, 1891.) F. H. P.